Transcultural Understanding of a Hereditary Disorder

Mucopolysaccharidosis VI in a Vietnamese Family

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A case study of a family referred for clarification of cultural issues illustrates how a transcultural psychiatric service developed in the pediatric hospital setting can be used to advantage. A Vietnamese family with an inherited disorder, Mucopolysaccharidosis type VI (Maroteaux-Lamy syndrome, MPS VI),[1] resisted genetic counseling and contraception. Three out of their six children were affected, one with a fatal outcome. The transcultural consultation offered an understanding of their behavior and facilitated subsequent management.

IN CLINICAL SETTINGS, medical anthropologists apply new theories and methods to understand how culture shapes the patient's understanding of his illness.[2–5] Clinicians have recognized the necessity of exploring what is behind the presenting problem, the "hidden agenda".[6–9] The importance of listening for the patient's understanding of illness and his "hidden agenda" is nowhere seen more clearly than in transcultural medicine.

In a culturally diverse setting, it is essential to have culturally appropriate medical, including pediatric, care. Although transcultural psychiatry has grown in general medicine,[10] these developments have not been extended fully to pediatrics. The work reported here stems from a new initiative in which a transcultural approach is applied in a major children's teaching hospital. In 1986 one of the authors, (IME) a child psychiatrist/medical anthropologist, initiated a pilot clinical service for cultural consultations. This service entailed the coordinated efforts of mental health professionals trained in medical anthropology and anthropologists with expertise in both Southeast Asian culture and language, and members of the refugee communities with professional backgrounds. In these consultations, understanding illness in a cultural context was of paramount importance.

Case Report

A family escaped from Vietnam in early 1982. They came from a coastal village in central Vietnam, where the father was a fisherman and the mother stayed at home with her two children. They spent 19 months in a Hong Kong refugee camp where a third child was born. Eventually they were accepted for resettlement in Australia, arriving in 1983.

Soon after, the mother conceived her fourth child, a girl. She was apparently well initially, but at 12 months, presented with a 2-week history of increasing dyspnea, cyano-

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sis, and lethargy. When admitted she was noted to be in severe congestive cardiac failure. Both her liver and spleen were palpable. On echocardiography,[1] she had dilated, poorly contracting ventricles. A full blood examination revealed marked granularity of the neutrophils suggestive of MPS VI. Despite intensive treatment, the infant died soon after admission. Her urine subsequently confirmed excessive dermatan sulphate excretion, and lysosomal enzyme studies on post mortem tissue showed undetectable levels of galactosamine-4-sulphate activity confirming MPS VI.

Shortly after the death of this infant, a fifth child was born. The family was requested to bring him for examination. However, he was not brought to this hospital until the age of 3 months, when he had developed severe heart failure. The findings were similar to those of his older sibling, but he responded well to medical therapy. Investigations again showed MPS VI. He has remained well on treatment but is showing increasing dysmorphic features consistent with MPS VI.

A pilot clinical service of mental health professionals and anthropologists for trans-cultural [illegible]s has been initiat[illegible]

Soon after the birth of the fifth infant the mother became pregnant again, being unable to tolerate the contraceptive pill. This infant was referred to the hospital at the age of 2 weeks because of the development of a murmur and tachypnea. Investigation revealed a moderately large patent ductus arteriosis which was ligated forthwith. Over the next few months, however, she developed increasing respiratory distress following a bronchiolitic-like illness. Despite the maintenance of reasonably good cardiac function she has required repeated admissions with cough, wheeze, and tachypnea. She has failed to thrive and her development is delayed. She too has developed dysmorphic features consistent with MPS VI.

The mother is currently pregnant and again did not accept the recommendation for amniocentesis, only seeking obstetric care late in her pregnancy. On this occasion, however, she seems reconciled to the idea of a tubal ligation.

Discussion

Understanding of Diagnosis

When patients are presented with diagnoses, they may already have their own explanations for their disorder or disease. Sometimes the patient integrates the physician's diagnosis with his own, and other times maintains both explanations simultaneously, not considering them mutually exclusive. Alternative explanations for disease often occur to patients from cultures different from our own as well as from our own culture. All patients have their own ways of understanding and explaining their illnesses, some analogous to those of biomedicine, some not. One must identify the patient's interpretation of the physician's explanation and also explore their non-biomedical beliefs about the disease so as to be helpful to the patient.[8]

Although counseled on several occasions, the father saw MPS, an autosomal recessive disease, as inherited solely from his wife's side. Both parents interpreted the disorder as inherited from the mother's sister because her sister died from "heart problems". The parents thought MPS was inherited directly from the mother's sister; the concept of genes was not important to their explanations.

Half of the children in this family have been afflicted with MPS VI; twice the predicted frequency. The father's response to an offer of further clarification of the genetic basis of MPS IV revealed what was important to him: "No, it would not change the fact that my daughter died."

The process of translating and explaining a word such as "hereditary" should not discourage the physician from pursuing the patient's or parents' understanding of the meaning of the translated word.

Understanding families' beliefs makes it possible to intervene more effectively.

The Family's Explanations

Patients can and often do explain their illnesses in a variety of ways. Southeast Asian patients draw upon Buddhism, indigenous folk beliefs, culture, and biomedical concepts introduced through exposure to Western medicine. According to Buddhist doctrine, one's status in the present life is determined by one's accumulated merit or demerit from previous lives.[11] Belief in *karma* can make the task of providing genetic counseling challenging. The parents, both Buddhist, believed that their childrens' illnesses were caused by destiny. The father said he believed in destiny and the logical cycle of life which repeats itself, as well as in the "Phong" (wind) theory of illness.

Although *karma* may explain the overall misfortune of a child or a family, more immediate causes founded in folk beliefs and folk medicine, such as malevolent spirits or humoral imbalances, are often cited for the etiology of illnesses.[12] "Wind illness"

explains a variety of symptoms and is one of the most common explanations for illness used by Vietnamese and other Southeast Asians.[13]

The family members were very concerned with the temperature extremes and "wind" of the small flat in which they lived as well as the cold and "wind" of Melbourne. They cited the harsh weather as a possible cause of their children's ill health. Their concerns about cold temperatures and especially "wind" were essential to understanding their explanations of etiology. These concerns were so important to them that they shifted homes despite enormous financial constraints.

Throughout much of Southeast Asia, both Chinese based astrology and indigenous variations of it have been used to explain and predict health and illness. Families may attribute astrological significance to certain illnesses, especially when such illnesses occur on corresponding dates (in this case the births of the sick children corresponded with the month in which one child died), or when ill family members are born under the same astrological sign.

Just as Western patients often seek alternative medicine, migrant patients have their own alternative medicine within their communities. Many Southeast Asian refugees remember the success of their own traditional healers. Patients may see the medical system here as inadequate and powerless for certain medical problems. This view is reinforced when Western medicine fails. The parents felt that their child died because "the hospital was not good enough and the doctors did not understand the medical problem."

Genetic Counseling

Genetic counseling often raises issues patients may not reveal to their doctors. When working with refugee and migrant patients these issues are particularly important to consider. An important issue for this family, not apparent at the time of counseling, was the importance of male offspring for ancestor worship.

It is essential to consider the nature of lineage descent and ancestor worship in order to understand why male children are of special importance. In parts of Vietnam it is believed that immortality depends upon an undying lineage. After parents die, their spirits must be properly honored by male descendants in order to preserve the family name and to attain reincarnation.[14,15] The father stressed the importance of having a son in Vietnamese culture. He explained that this was why he and his wife became very upset when they found out their only son was also ill. The role of a son in the reincarnation of parents most likely explains their wish to have more children in the hope of having a healthy son, a wish not verbalized.

Patients who are not listened to or are told they are wrong will not reveal their belief systems about illness.

Conclusion

Exploring a patient's concept of illness requires some knowledge about the patient's belief systems. In the case discussed, predestination, reincarnation, male lineage, and "wind illness" were particularly important. However, as clinicians cannot possibly have the necessary background knowledge for all their patients, they must enquire about the relevant beliefs. Patients who are not listened to and who expect to be ridiculed for their beliefs or told that their beliefs are simply wrong will not reveal their belief systems about illness. Understanding of these beliefs make it possible to intervene more effectively to prevent further hardship to the family and to relieve much of the distress experienced.

Genetic counseling for migrant and refugee patients requires an exploration of the cultural significance of children, lineage, and kinship structure. Just as it is important to work with any patient's beliefs about the etiology of his illness, it is important to explore and incorporate into treatment culturally relevant issues for genetic counseling. The patient's view of his illness will enhance his compliance and satisfaction. Through treatment of migrant patients, whose values and beliefs may differ drastically from those of their care-givers, one may learn the importance of exploring these relevant values and beliefs for all patients counseled.

Further research is needed to help understand the impact of cultural factors on families whose children suffer severe disorders. Physicians faced with such devastating inherited disorders like MPS IV naturally want to influence patient behavior. This case has shown why the best of clinical intentions can fall short. It also demonstrates how a transcultural liaison service of the type initiated at the Royal Children's

Hospital can help to make health care more culturally appropriate and subsequently more effective, while providing the opportunity for important clinical research.

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